

CASE REPORTS

Ebstein's anomaly associated with splenomegaly and reversible hypersplenism

M R Bennett, M F Shiu

Abstract

A case of Ebstein's anomaly associated with chronic right heart failure, hepatosplenomegaly, and the haematological features of hypersplenism is presented. The haematological abnormalities were corrected after tricuspid valve replacement but recurred with the re-emergence of clinical features of right heart failure.

Ebstein's anomaly often results in right heart failure with an increase in systemic venous pressures. The latter, when transmitted as raised portal venous pressures, can cause splenomegaly and the haematological features of hypersplenism. The occurrence of reversible hypersplenism associated with changes in right sided pressures in Ebstein's anomaly has not been described before.

Case report

Ebstein's anomaly was diagnosed in a child with cyanosis on exertion, characteristic physical findings, and radiological evidence of cardiomegaly. He remained well, however, without treatment, until the age of 35 when he presented with dyspnoea and epistaxis. On examination he had cyanosis at rest, features of right heart failure with tricuspid regurgitation, hepatosplenomegaly, and widespread purpura. Investigations performed at that time showed a haemoglobin concentration of 188 g/l (normal range 135-165), white cell count of $3.0 \times 10^9/l$ (normal range $4.0-11.0 \times 10^9/l$), and a platelet count of $75 \times 10^9/l$ (normal range $150-450 \times 10^9/l$). Cardiac catheterisation showed raised right sided pressures and tricuspid regurgitation (table) and normal left sided pressures. At operation, undertaken on the basis of symptoms and evidence of heart failure, the tricuspid valve was shown to be incompetent with an aneurysmally dilated right atrium, small right ventricle, and a patent foramen ovale. The

tricuspid valve insertion was typical of Ebstein's anomaly. Initial surgical intervention was plication of the right atrium, which made the tricuspid valve competent, together with closure of the foramen ovale. However, there was little improvement in symptoms after this operation. Right heart failure became worse over the next two years and he required further admissions. Investigations at this time showed a haemoglobin concentration of 175 g/l, a fall in his white cell count to $2.2 \times 10^9/l$, and a platelet count of $70 \times 10^9/l$. Cardiac catheterisation showed a further increase in his right sided pressures (table) and another operation was considered.

The tricuspid valve was replaced with a Starr-Edwards prosthesis two years after valvoplasty and his clinical signs of right heart failure resolved with a considerable improvement in symptoms. This was associated with a dramatic reduction in his hepatosplenomegaly as assessed clinically and on abdominal radiography and a sharp rise in his neutrophil and platelet counts, which restored them to normal (figure). Between 1978 and 1989 signs of right heart failure gradually re-emerged with progressive hepatosplenomegaly and concomitant falls in his neutrophil and platelet counts (figure). Cardiac catheterisation showed a further increase in right sided pressures (table).

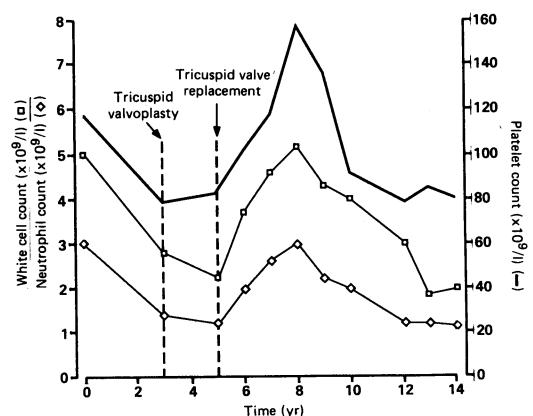
At each admission the patient was investigated for causes of his pancytopenia. Bone marrow samples showed a consistently hypercellular marrow, with increases in all cell

Department of Cardiology, The Queen Elizabeth Hospital, Birmingham
M R Bennett
M F Shiu

Correspondence to Dr M R Bennett, Department of Cardiology, University of Wales College of Medicine, Heath Park, Cardiff CF4 4XN.

Pressure recordings (systolic/diastolic (mm Hg)) at cardiac catheterisation before each operation and 16 years after replacement of the tricuspid valve

Chamber	Before valvoplasty	Before tricuspid valve replacement	16 years after tricuspid valve replacement
Right atrium	11/1 (mean = 6)	30/15 (mean = 24)	33/20 (mean = 27)
Right ventricle	22/0 (mean = 12)	30/15 (mean = 23)	57/20 (mean = 39)



White cell, neutrophil, and platelet counts during each operation and at follow up. (Means of three observations.)

lines, but no evidence of megaloblastosis, dysplasia, or infiltration. Intravascular haemolysis was excluded by a normal peripheral blood film, serum haptoglobin, and urinary haemosiderin. Coombs test and the Ham acid lysis test were negative and serum vitamin B₁₂ and folate concentrations were normal. Anti-DNA antibodies and the rheumatoid arthritis latex test were negative. Infective endocarditis was excluded by repeated negative blood cultures. The pancytopenia was therefore attributed to hypersplenism. An ultrasound scan of the abdomen confirmed hepatic venous congestion and hepatosplenomegaly, and the presence of splenic varices implied portal hypertension, though the portal vein was patent and of normal size.

Discussion

Hypersplenism is defined as a depression of one or more of the cell counts in the circulating blood that can be wholly attributed to splenic enlargement and is associated with a cellular bone marrow.¹ Other causes of pancytopenia: hypersplenism, marrow aplasia/dysplasia, marrow infiltration, megaloblastosis, systemic lupus erythematosus, and paroxysmal nocturnal haemoglobinuria were excluded as indicated above. The patient's initial polycythaemia is usual in cases of Ebstein's anomaly with right to left shunt,² and was corrected by closure of the shunt.

Our patient with Ebstein's anomaly had had chronic right heart failure with raised systemic venous pressures for many years. Chronic right heart failure with hepatic venous congestion is associated with splenomegaly in 12–25% of patients^{3–5}; the mechanism is believed to be portal venous hypertension secondary to raised hepatic ven-

ous pressures. Moderate to severe symptoms in Ebstein's anomaly or the presence of heart failure are regarded as indications for tricuspid valve plication or replacement.⁶ Replacement has been shown to reduce right atrial pressures,⁶ and should therefore reduce both systemic and portal venous pressures. Reduction in portal venous pressures, for example after distal spleno-renal shunting procedures, reversed the neutropenia and thrombocytopenia of hypersplenism and this was associated with a reduction in splenic size.^{7,8}

Hypersplenism in our patient seems to be the result of raised systemic (and thus portal) venous pressures, and treatment of the cardiac lesion with reduction in these pressures improved the secondary haematological characteristics. Such an association between Ebstein's anomaly and hypersplenism has not been described before; perhaps because detailed longitudinal studies over many years are often not possible. Our patient is also unusual because he has survived 17 years after the onset of heart failure—a poor prognostic sign in patients with Ebstein's anomaly.

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